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Surgical Treatment of Sacrococcygeal Chordoma

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Background: Sacrococcygeal chordomas are difficult to manage.
Methods: The treatment results of 12 patients with sacral chordomas were analyzed. Four patients had local relapse before they were referred to our hospital: three patients previously underwent two surgeries, and one patient one surgery and irradiation (60 Gy). In this institute, 10 patients underwent surgery alone, and one had surgery plus postoperative irradiation. One patient underwent radiotherapy alone due to an inoperable lesion that reached to the L5 spine. For seven surviving patients, the average follow-up period was 50 months.
Results: One of four patients with a marginal margin and one of two patients with a contaminated margin developed local relapse. Of five patients with intralesional margins, four patients developed local relapse and two had metastasis to the spine and lungs 36 and 15 months after surgery, respectively. One patient with irradiation alone had metastasis to the lungs 15 months after initiation of treatment. Three patients died due to progression of the disease, one of a heart problem, and one of apoplexy. One of 10 patients with implantation of gentamycin beads after removal of tumor, developed infection.
Conclusions: For local control of sacral chordomas, an adequate surgical margin is important. Gentamycin beads may be effective to control postoperative infection of the dead space.
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KEY WORDS: **bone; neoplasms; chordoma; surgery; complication**

INTRODUCTION: SURGICAL COMPLICATIONS

Chordoma is a malignant tumor arising from remnants of the notochord [1]. Approximately 50% of chordomas occur in the sacrococcygeal region [2,3]. The tumors occur twice as frequently in men as in women and are uncommon under the age of 40 years [4]. The large vacuolated cells (physaliferous cells) are a characteristic of this tumor [5]. Chordoma is considered to be of low-grade malignancy, but surgery with a safe margin is difficult, mainly due to anatomical characteristics of the sacrum. Moreover, many patients with sacral chordoma have been previously treated inadequately [2]. These inadequate treatments finally make adequate surgery more difficult and lead to a poorer prognosis of the patients. The surgical complications, including neurological impairment, also become serious problems in patients with a sacral chordoma. In our hospital, we have been using gentamycin beads to prevent infection in the dead space after removal of large sacral tumors. We retrospectively analyzed the treatment outcome of 12 patients with a sacral chordoma. We also report the clinical and radiographic features and the results of the prophylactic treatment used to minimize infection in patients with sacral chordoma.

MATERIALS AND METHODS

Between 1982 and 1993, 12 patients who had a sacral chordoma were treated in the Department of Orthopae-

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dics of the Westfälishe Wilhelms-University, in Münster, Germany (Table I). Four patients had local relapse before they were referred to our hospital. Three patients previously underwent two surgeries and one patient one surgery and irradiation (60 Gy). The medical records, histologic sections, and radiographs were reviewed for each patient. Information regarding tumor stage, tumor extension, and treatment outcome were available for all patients.

The patients reported that the symptoms had been present for a period of 6 months to 3 years before the initial diagnosis was made (average 1.2 years). Eleven patients had had pain in the sacrococcygeal region, sometimes radiating to the buttocks or legs. One patient had constipation and urinary incontinence rather than pain.

The subjects were 11 male patients and one female patient, between 52 and 80 (average 62.5) years old. The tumor volume was evaluated according to the method described by Göbel et al. [6]. The size ranged between 81 and 1755 ml (average 519 ml). Preoperative evaluation of the surgical stage included plain radiographs, bone scan, CT scans, and MR imagings. Surgical stage was evaluated according to Enneking's criteria [7]: IA; 2, IB; 10.

Eleven patients underwent a surgical resection of the tumor. All surgical margins were classified according to Enneking's criteria [7]. The highest level of tumor infiltration was L5 in one, S1 in 2, S2 in 3, S3 in 5, and S4 in one. The proximal extent for the posterior approach was S3 in 3 and S4 in one patient. The proximal extent for the combined anterior and posterior approach was S1 in 2, S2 in 3, and S3 in one patient. One patient (Case 6) needed irradiation of 55 Gy after an intralesional surgery in the judgment of the doctor in charge. One patient (Case 10) received irradiation alone because he was diagnosed as having an inoperative lesion due to extension up to the 5th lumbar spine. When the most caudal preserved nerve was S1, colostomy was performed except in Case 9. When both S2 nerves or lower were preserved, colostomy was not performed except in Case 4. Five patients underwent colostomy as a part of the index operation in this institute. The dose of irradiation in the patient who received irradiation alone was composed of 16 MeV photons (total doses: 41.6 Gy, daily doses: 1.6 Gy) and 14 MeV neutron (total doses: 7.2 Gy, daily doses: 0.6 Gy).

After removal of the tumors, gentamycin beads (Septopal-10er-Kette, Merck, Darmstadt, Germany) were implanted in 11 patients and the cavity was covered by a gluteus maximus flap (Fig. 1). At 3 months following implantation, all the beads were removed.

Clinical follow-up was done every 3 months, the chest roentgenogram was checked every 6 months, computed tomography or magnetic resonance imagings of the pelvis was done every 3 months. If necessary at the time of the follow-up, data were gathered by means of a questionnaire or telephone interview from the patients' family or doctors who observed the patients.

RESULTS

For the seven surviving patients, the mean follow-up was 50 months (range 24–90 months). Overall, at latest follow-up, six patients showed no evidence of disease, one was alive with local disease, three had died of disease, and two had died of complications. Death from complications or disease occurred at a mean of 22 (range 10–45) months.

Four patients had undergone previous surgical treatment. Case 1 had local relapse 12 and 6 months after the first and second previous surgeries, respectively. Case 2 had a local relapse 22 months after the previous surgery and received irradiation. Case 6 had local relapses 36 and 48 months after the first and second surgeries, respectively. Case 9 had local relapses 12 months after the first, and again 12 months after the second surgery. All these surgical procedures were done with an intralesional margin from the posterior approach.

As for surgery in this institution, four patients had surgery with a marginal margin and two had contaminations of the margins. Five patients had surgery with intralesional margins. The duration of surgery by posterior approach ranged between 3.4 and 7.5 hours (average, 4.9 hours) and that by anterior-posterior approach ranged between 9.5 and 21.0 hours (average, 12.5 hours). The blood loss from posterior surgery ranged between 3,100 and 8,500 ml (average 4,680 ml) and from anterior-posterior surgery, between 2,100 and 28,800 ml (average 9,833 ml).

Six patients developed local relapses (4 of 5 intralesional, one of 4 marginal, and one of 2 contaminated margins). Although one patient treated with adjuvant radiotherapy following intralesional excision (Case 6) has not shown local tumor progression, the chemotherapy proved ineffective, and he had spine metastasis 36 months after a previous surgery. Four patients underwent further resection of the locally relapsed lesion following our index procedure. One (Case 2) of four patients developed local relapse again and is still alive with the disease. Three patients (Cases 4, 5, and 11) are alive with no sign of the disease. Two patients (Cases 1 and 3) died due to progression of the disease 5 and 12 months, respectively, after local relapse. Three patients had metastasis to the thoracic spine and the lungs during the follow-up after surgery by us. After spinal metastasis, Case 6 had paraplegia below the Th7 level. He underwent laminectomy, posterior spinal fusion with instrumentation, and postoperative irradiation of 30 Gy. Finally, he developed multiple metastasis and died. One patient (Case 10) died of concurrent disease (apoplexy) 7 months after the appearance of pulmonary metastasis. The other patient (Case 3) died due to a lung metastasis.

TABLE I. Characteristics, Treatment, and Outcome of Patients with Sacral Chordoma, Treated at the Department of Orthopaedics of the Westfälische Wilhelms-University, Münster, Germany

				Surgery in this hospital									
Patient no.	Age/sex	Size (ml)	Previous treatment (times)	Surgery (surgical margin)[a]	Time (hr)	Blood loss (ml)	Most caudally preserved nerve	Colostomy[b]	Gentamycin beads[c]	Adjuvant treatment[d]	Complications and postoperative status	Outcome[e]	Follow-up (mos)
1	52/M	178	Surgery (2)	Posterior (IL)	7.5	8,500	S2–S2	−	+	—	Bowel and bladder dysfunction, sexual problems, local relapse (5 mo)	DOD	10
2	64/M	1,755	Surgery (1) Rad : 60 Gy	Anterior + posterior (IL)	21.0	6,700	S1–S1	+	+	—	Hematoma, pneumonia, bladder dysfunction, sexual problems, local relapse (18 mo)—resection, local relapse (24 mo)	AWD	36
3	72/M	93	—	Posterior (IL)	4.0	4,000	S2–S3	(+)	+	—	Renal failure, heart problem, bladder dysfunction, local relapse (12 mo), lung metastasis (15 mo)	DOD	24
4	56/M	374	—	Anterior + posterior (M)	10.5	3,600	S3–S2	+	+	—	Skin necrosis, local relapse (72 mo)—resection (M)	NED	90
5	64/M	446	—	Anterior + posterior (IL)	9.5	2,100	S1–S1	−	+	—	Bowel and bladder dysfunction, sexual problems, local relapse (72 mo)—resection (IL)	NED	90
6	60/M	360	Surgery (2)	Posterior (IL)	6.5	4,500	S4–S4	−	−	Chemotherapy Rad 55 Gy	Spine metastasis (36 mo)—laminectomy, Rad (30 Gy)	DOD	45
7	59/M	81	—	Posterior (M)	3.4	3,100	S3–S3	−	+	−	Temporary bowel and bladder dysfunction	CDF	43
8	55/M	624	—	Anterior + posterior (M)	11.3	28,800	S1–S1	+	+	−	Bladder dysfunction, sexual problems	CDF	40
9	76/F	899	Surgery (2)	Anterior + posterior (M,C)	12.0	5,800	S1–S1	(+)	+	—	Skin necrosis, hip disarticulation, hematoma perianal infection, decubitus, bladder dysfunction, heart failure	DOC	11
10	60/M	167	—	—	—	—	—	—	—	Rad. 48.8 Gy	Pulmonary metastasis (15 mo), apoplexy	DOC	22
11	53/M	714	Transrectal biopsy	Anterior + posterior (M,C)	10.5	12,000	S1–S1	+	+	—	Bladder dysfunction, sexual problems, local relapse (7 mo)—resection (M, C)	NED	24
12	80/M	537	—	Posterior (M)	3.5	3,300	S2–S2	—	+	—	Bowel and bladder dysfunction, bladder, skin fistula	CDF	24

[a]IL: intralesional margin, M: marginal margin, C: contaminated margin.
[b]+: performed, −: not performed, (+): performed later.
[c]+: used, −: not used.
[d]–: not performed.
[e]DOD: dead of disease, AWD: alive with disease, NED: no evidence of disease, CDF: continuous disease-free, DOC: dead of complication.

Fig. 1. Case 12. **A.** Preoperative X-ray. Destruction of the lower part of the sacrum is noted. **B.** Postoperative X-ray. Gentamycin beads were put in the dead space after removal of the tumor.

Three patients continued to be disease-free at the mean follow-up period of 36 (24–43) months.

Bowel or urinary disturbances appeared in nine patients. Of the five patients whose bilateral S1 was the lowest preserved nerve, four patients underwent colostomy as an index operation and one underwent colostomy later, due to impaired bowel control. All five patients had permanent dysfunction of the bladder, and one patient had impaired bowel control. Both patients whose bilateral S2 was the lowest preserved nerve had permanent dysfunction of the bladder and bowel. Of the two patients whose preserved nerves were S2-S3, one had permanent and the other temporary dysfunction of the bladder. One had undergone colostomy as an index surgery and the other had colostomy later due to impaired bowel control. One patient whose lowest preserved nerves was S3-S3 had a temporary dysfunction of the bowel and the bladder. One patient with both S4 intact had no bladder or bowel problems. Nine patients who had an impaired bladder function required chronic intermittent bladder catheterization. Two patients underwent colostomy a few months later due to impaired bowel control.

The complication rate excluding oncological and neurological problems was 50% (6/12). One had pneumonia (Case 2), one chronic renal failure due to hypertension (Case 3), two heart failure (Cases 3 and 9), one deep infection (Case 9), one decubitus (Case 9), one apoplexy (Case 10), and one bladder-skin fistula (Case 12). Four patients had local wound problems, two with skin necrosis (Cases 4 and 9) and two with hematoma (Cases 2 and 9). One skin necrosis (Case 4) healed following simple suture after partial resection of the necrotic skin. The necrotic lesion of the other patient (Case 9) was too large to be healed by additional surgery of skin revision, and she underwent hip disarticulation due to a large necrosis from the pelvic to hip region. In these patients, the free-flap was not used. The Case 9 patient also had a perianal infection with ‘‘decubitus,’’ and these disorders were not cured at the time of her death. Two hematomas without infection were healed by revision surgery with curettage. One patient had a heart problem and died without relapse (Case 9). One patient died of apoplexy after pulmonary metastasis as previously mentioned.

DISCUSSION

Sacral chordoma is a low-grade malignant tumor, but it is locally aggressive [1]. After surgery of chordoma, relapse and complications frequently occur [8]. It is one of the most difficult tumors for the orthopaedic surgeon, a frequent complication being infection. To solve this problem, we used implantation of gentamycin beads in the big dead space after removal of the tumor.

There have been several reports on the treatment outcome of sacral chordomas [4]. In 20 patients with sacral chordomas treated at the Memorial Sloan Kettering Cancer Center (New York City) [9], eight patients had additionally received irradiation and two chemotherapy as an adjuvant treatment. Kaiser et al. [10] reported the treatment outcome of 63 patients. There was no information

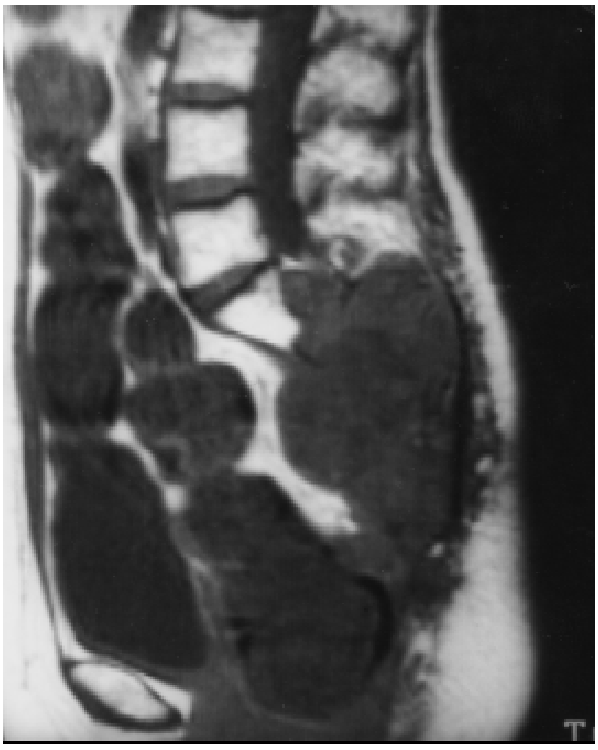

Fig. 2. Case 9. T1-weighted magnetic resonance image of the sagittal plane. The low signal intensity tumor mass infiltrated in the upper part of the sacrum with producing an extraskeletal tumor component.

about adjuvant treatment, but 55 patients had posterior excision, seven an incisional biopsy, and one had anterior resection. Samson et al. [11] recently reported the treatment results after posterior surgery; the patients who had been previously treated with excluded and 76% of the patients received radiation therapy. Stephens and Schwartz [12] reported six cases of sacrococcygeal chordomas operated on by the combined anterior and posterior approaches. Three of the six received irradiation.

Sacral chordomas often become very large before correctly diagnosed. An average interval between the onset of symptoms and correct diagnosis was as long as 1.2 years in this series. After computed tomography and/or magnetic resonance imaging was used for diagnosis, the tumor of the sacrum was identified in all cases in this series (Fig. 2). However, previous surgery had been performed inadequately in four patients. One patient (Case 11) had undergone transrectal biopsy that should not be performed for making histologic diagnosis because the periosteum of the sacrum and the presacral fascia works as a barrier against the tumor extension [8]. Twenty-nine of the 52 patients treated at Massachusetts General Hospital (Boston) had had a primary operation elsewhere and had been referred after at least one local recurrence [11]. Our finding of a median interval of over 1 year between onset of symptoms and correct diagnosis is typical of previous reported series [5,9].

There are several approaches to the sacrum [8,13,14–16]. In tumors with osseous involvement below the third sacral segment, the posterior approach is recommended [3,9]. For lesions extending to the S1 or S2 body, a combined anterior and posterior approach is indicated [11,15,17–19]. However, since Stener and Guntherberg [8] suggested the possibility of an abdominal hernia occurring in previously treated cases, we used a retroperitoneal approach with two separate incisions through the skin along the lateral border of the aponeurotic sheath of the rectus abdominus in this series [20]. As the rectus abdominus muscles remain intact and functional, in this approach they continue to help empty the bladder and bowel regularly [20]. Tomita and Tsuchiya [16] reported three patients who underwent total sacretomy without colostomy. Their patients controlled their bowels by using enemas. However, we prefer colectomy, which allows elderly patients to live without using diapers and improves their quality of life.

In this series, the local relapse rate after intralesional surgery was 60% and local relapse rate after surgery with marginal margin without contamination was 25%. Kaiser et al. [10] reported a 64% recurrence rate after tumor spillage and 28% recurrence rate without tumor spillage. Of the 20 patients with sacral chordomas reported by Sundaresan et al. [9], 60% had local relapse and 30% metastasis. According to Samson et al. [11], the disease-free survival rate at 10 years in 21 patients treated by posterior surgery was 60%. However, they excluded the patients who had been previously treated.

Metastasis developed in 3 of 12 patients in this series. The reported rates of occurrence of metastases range from 5% to >40% [1,3,10,11,21,22]. However, in the treatment of sacral chordoma, death not only due to tumor progression, but also due to complications is also important. When the local resection is incomplete, recurrences would sometimes develop at between 1 and 6 years after surgery; this was different from the report by Sundaresan that recurrence developed within the first few years [3].

Bladder and rectal function after surgery were major problems. Bowel or urinary disturbances appeared in 9 of the 12 patients in this series. They are directly related to the number of preserved nerve roots [8]. If the bilateral S2 nerve roots are sectioned, normal urogenital and rectal functions are lost [3,9]. If both S2 nerve roots can be preserved, 50% of the patients can regain at least partial bladder and bowel control [11]. It is important to remove as much of the lesion as possible and spare as many nerve roots as feasible [11]. However, the surgical margin should not be compromised because additional surgery with adequate margins after local relapses becomes more difficult and more nerve roots must be resected.

Infection and delayed wound healing often complicate the postoperative course [8,20]. According to the report

by Samson et al. [11], 33% of the patients had wound problems. Since the infection rate was high in our previous experience, we filled the cavity with a bag of gentamycin beads [20] and covered it with a gluteus maximus flap. Moreover, we inserted a drainage until the total daily discharge became <20 ml. In this way, only one of 11 patients who underwent surgery developed an infection in the operative region. The gentamycin beads were removed one to several months later to lessen the risk of foreign body reactions. At the removal of the beads, the cavity was always filled with healthy granulation tissue.

There were several serious complications (pneumonia, renal failure, heart failure, apoplexy). A bladder-skin fistula was also noted. Most of the patients are elderly and they may have had a past history of chronic diseases. Surgery tends to be extensive and time-consuming, so many complications must be expected and taken care of in advance and during the follow-up period. The cure rate by surgery always must be compared with serious complication rates accompany surgery. Palliative surgery may be justified in the elderly patients. Recently, good local control of chordoma after neutron beam therapy has been reported [23]. If it is available, this type of irradiation may be encouraged in patients with inoperable lesions, those who refuse major surgery, or in patients with the risk of a high complication rate.

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